Papilloedema and visual failure in a patient with nocturnal hyperventilation

A morbidly obese woman treated for polymyositis developed symptoms of raised intracranial pressure and visual failure. Invasive CSF pressure monitoring showed normal pulsatile pressure but regular nocturnal episodes of raised intracranial pressure during sleep. These episodes were associated with hypoxia and hypercarbia, and suggested a possible role of hypercarbia in the development of raised intracranial pressure, visual failure, and nocturnal hyperventilation. A 45 year old woman was admitted for investigation of headaches, papilloedema, and progressive visual failure. She was morbidly obese (weight 126 kg, body mass index 52.7 kg/m²) and gave a history of recurrent deep vein thrombosis, pulmonary emboli, and polymyositis (treated with prednisolone and azathioprine). Examination showed bilateral papilloedema with visual acuities of 6/24 in the right eye, and 6/18 in the left eye. A bulbar papillae had been only successful in the sitting position; hence an accurate measurement of the CSF opening pressure had not been recorded. Biochemical and cytological analysis of the CSF showed it was unremarkable, and CT of the head was normal. A clinical diagnosis of benign intracranial hypertension was made and a neuroophthalmological opinion sought. A left optic nerve fenestration was performed without improvement, and so detailed assessment of the intracranial pressure was requested. Access to CSF was achieved with a right frontal ventricle catheter connected to a subcutaneous reservoir. Continuous monitoring of intracranial pressure was undertaken from the reservoir (Camino optical transducer) through a fluid filled transcutaneous butterfly needle (21G). Radial arterial blood pressure, middle cerebral artery flow velocities by transcranial Doppler (Scimed), and continuous measurements of peripheral oxygen saturation by MultiNex oximetry were also taken, and displayed graphically on a portable computer. The cerebral perfusion pressure (mean radial arterial blood pressure-mean intracranial pressure) and pulsatility of the middle cerebral artery flow velocity (FV) were calculated (pulsatility index P = FV amplitude/FV mean) were calculated. Recordings were carried out during the night with the patient in her usual sleeping position (20 degrees head up) for up to 10 hours. Before sleep, baseline intracranial pressures of 5-15 mm Hg were recorded. Within one hour of sleep, cycles of high pressure waves (40-45 mm Hg) lasting 10-20 minutes were evident, occurring every 60-120 minutes and superimposed on higher background pressures of 15-25 mm Hg (fig 1). The intracranial pressure waves were accompanied by a fall in cerebral perfusion pressure to as low as 40 mm Hg, and were tightly coupled with increases of middle cerebral artery flow velocity and decreases in cerebrovascular resistance. Respiration function was evaluated further; arterial blood gases showed daytime hypoxia (mean PaO₂ 65 mm Hg with normal Pco₂ 35 mm Hg). Overnight ventilation studies indicated a mean baseline arterial saturation of 82-8% and end tidal Pco₂ of 6.1 kPa. During the periods of raised intracranial pressure,
cerebral vasodilatation. We therefore suggest that nocturnal hyperventilation producing increased intracranial pressure secondary to increased cerebral blood volume contributed to the symptoms. Chronic respiratory disease with severe hypercapnia has long been regarded as a cause of raised intracranial pressure and papilloedema but there were no such features in this case, and although a significantly raised CSF pressure is required for the diagnosis of benign intracranial hypertension, the clinical and radiological features in our patient were typical of this condition. Further, low baseline CSF pressures are often found in patients with chronic benign intracranial hypertension despite persisting papilloedema. Two important points are raised. Firstly, abnormal CSF dynamics require continued observation over several hours as baseline CSF pressure may be normal and waves of raised intracranial pressure transient. Inadequate attention to CSF dynamics may partly explain why isolated CSF pressure estimations do not predict the development of papilloedema and visual deterioration. Secondly, although nocturnal hyperventilation has not been quoted as a contributing factor in benign intracranial hypertension, a relation with raised intracranial pressure has been found. Overnight monitoring of peripheral oxygen saturation may be a useful addition to the investigation of obese patients with symptoms of raised intracranial pressure.

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Hemichorea reversible after operation in a boy with cavernous angioma in the head of the caudate nucleus

Hemichorea and hemiballismus is a structural lesion in the contralateral basal ganglia with a large list of possible causes, including various vascular malformations. Cavernous angiomas are congenital vascular malformations that are surrounded by cavernous angiomas include epilepsy, acute signs secondary to (recurrent) bleeding, and rarely progressive neurodegenerative disease due to expansion of a mass of haemosiderin within the angioma. With the availability of MRI the number of clinical reports on the subject of CVMs has increased. Recently a case was reported of cavernous angioma in the lentiform nucleus that was the first to present with a movement disorder, in this case focal dystonia. Complete resection was followed by resolution of the symptoms.

We report a 11 year old boy with cavernous angioma in the caudate nucleus, presenting with contralateral hemichorea, evidence of recurrent bleeding, and the disappearance of the hemichorea after surgery. The boy complained of involuntary movements of the right half of his body including his face, arm and leg, that had suddenly started the week before admission. He could not suppress these movements. There was no family history of neurological disease.

The neurological examination on admission showed continuous, random, jerking movements of the face and extremities on the right side of the body. Muscle strength, sensation, reflexes were normal.

Brain MRI (figure A) showed a lesion in the head of the caudate nucleus, with the typical aspect of a cavernoma.

Two weeks later the boy experienced a sudden deterioration, with involuntary movements of a larger amplitude, more appropriately termed hemiballismus. Surgery was considered appropriate.

With the Leksell stereotactic frame (Elekta Co, Sweden) the shortest route to the lesion via the paramedial frontal lobe was estimated. At the time that the surgery hole was made and a silastic tube was passed to the border of the lesion with a Backlund catheter implantation set. After craniootomy the lesion was reached with the catheter as a guide. The Muller bulb like vascular lesion was removed completely, including two small haemorrhages.

Histology (figure B) showed a conglomerate of vessels and hamartomatous tissue. The wall of these channels consisted of a single inner layer of endothelial cells and an outer layer of collagen of varying thickness. Some vascular spaces were occluded by a recent or an organised haemorrhage and some vessel walls were partly calcified. Iron pigment was found in and around several vessels, as evidence of prior bleeding. The surrounding brain tissue showed pronounced gliosis and presence of iron.

In the two months after the operation the hemichorea-hemiballismus disappeared completely. Control MRI (figure C) showed complete removal of the angioma.

This case is to our knowledge the first in the literature of a histologically confirmed cavernous angioma presenting with hemichorea. Hemichorea has been described in lesions of the caudate nucleus, and is thought to reflect release phenomena caused by a lesion of the striatal neurons projecting to the external globus pallidus.

The incidence of cavernous angiomas remains obscure. In a consecutive series of 11 children operated on for cerebral vascular malformations five were diagnosed to have cavernous angiomas. Scott et al. state that in some paediatric institutions cavernous angiomas are the most common cerebral vascular malformations encountered. Most cavernous angiomas, however,